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CASTLEMAN'S DISEASE IN THE PARARENAL RETROPERITONEAL SPACE, WHICH IS INDISTINGUISHABLE FROM RENAL CELL CARCINOMA : A CASE REPORT

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A 29-year-old woman was hospitalized in our institute with the diagnosis of a right renal mass by ultrasonography on medical checkup. Computerized tomography showed a lower pole solid mass (9 cm in diameter), which was enhanced homogeneously, as well as the renal cortex in the arterial phase. The tumor was excised using radical nephrectomy based on the preoperative diagnosis of renal cell carcinoma, and thus lymph node dissection was also performed. The excised tumor was isolated from the kidney in a thin capsule, macroscopically. Postoperative pathological diagnosis revealed hyaline vascular type Castleman's disease. There was no recurrence at 1 year after the operation without any adjuvant therapy because of the complete resection.

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Key words : Castleman's disease, Retroperitoneal space, Renal cell carcinoma

INTRODUCTION

Castleman's disease (CD) is a rare tumor of lymphoid tissues that was first reported by Castleman[1] . Approximately 70% of CD is usually generated in the mediastinum and only 7% forms in the retroperitoneal space[2–4]. In addition, Bucher et al. reported 97 cases of retroperitoneal CD, and showed that 25% (24 cases) occurred in the "pararenal space"[5]. Most cases of CD in the retroperitoneal space are diagnosed as malignant tumors such as renal cell carcinoma (RCC) preoperatively. Imaging techniques such as ultrasonography (US), computerized tomography (CT), magnetic resonance imaging (MRI) and angiography are not conclusive for diagnosis of CD, and confirmed diagnoses are usually based on pathological findings from the resected tumor. We report a case of CD located in the pararenal retroperitoneal space, which was indistinguishable from RCC on preoperative images.

CASE REPORT

A 29-year-old "healthy-looking" woman was hospitalized in our institute with a large tumor in the right kidney that had been identified in another hospital. She did not have any symptoms in an initial medical examination, whereas physical examination revealed a palpable abdominal mass on the right lateral lesion without tenderness. There were no inflammatory findings in a blood test (white blood cell count, 6,100/μl ; C-reactive protein, 0.09 mg/dl). However, three-phase dynamic CT showed a large mass in the lower pole of the

Fig. 1. Dynamic CT shows a solitary tumor with enhancement. In the arterial phase, the tumor was strongly enhanced as well as renal parenchyma. In contrast, in the venous phase, enhancement for the tumor was weaker than that of renal parenchyma. (A) : arterial phase, (B) : venous phase.

Fig. 2. T2-weighted retroperitoneal MRI shows a homogeneous tumor in the lower pole of the right kidney (arrow).

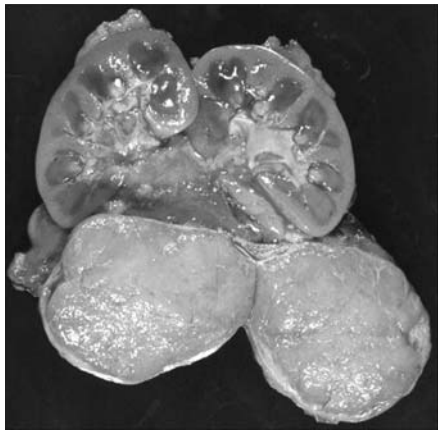

Fig. 3. Macroscopic findings demonstrate a lobulated tumor with a capsule. The tumor was located on the anterior side of the lower right kidney.

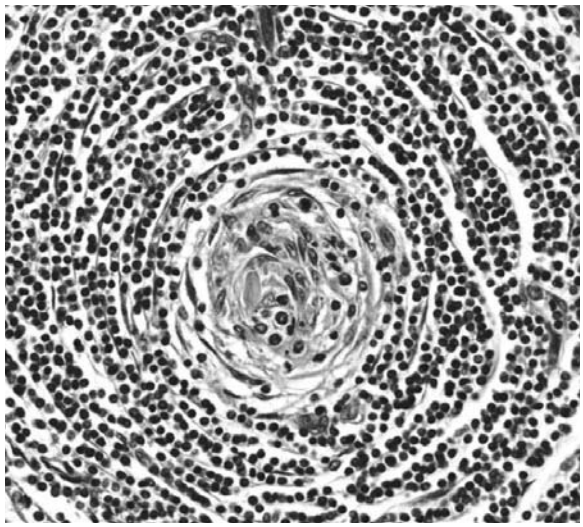

Fig. 4. Microscopic findings demonstrate small lymphoid follicles, which had marked vascular proliferation and hyalinization.

right kidney (size: approximately 10 cm in diameter) without internal calcification. The tumor was enhanced homogeneously in both the arterial and venous phases, and strong enhancement was observed in the arterial phase as well as renal parenchyma (Fig. 1A). In contrast, the venous phase enhancement was comparatively weaker than that of renal parenchyma (Fig. 1B). The findings of CT appeared to be that of typical RCC. MRI also revealed that the tumor was a solitary homogeneous mass (T1-weighted: low intensity and T2-weighted: high intensity) (Fig. 2). Lymph node swelling was not detected on either CT or MRI. Therefore, we made the diagnosis of RCC (clinical stage: T2N0M0), and then radical nephrectomy and lymph node dissection were performed.

The tumor was found on the right side of the vena cava through a median incision, after mobilization of the ascending colon and duodenum. The tumor was covered by the renal fascia, and we could not identify any plane for division between the tumor and kidney during the operation. Thus, the mass was excised in an en-bloc manner with a wide margin in pararenal fat. There was no evidence of lymphadenopathy. Total weight of the resected mass was 520 g. By macroscopic observation, the tumor was $7 \times 9 \times 5$ cm in size, was located on the anterior side of the lower portion of the right kidney, and was completely apart from the kidney with thin capsule. The tumor had a yellowish surface with granular findings (Fig. 3). Thus, we conclude that the tumor did not originate in the kidney and was derived from extra renal tissue.

Pathological examination of the resected tumor showed increased small lymphoid follicles. The follicles demonstrated marked vascular proliferation and hyalinization of their abdominal germinal centers (Fig. 4). Finally, diagnosis of this tumor was hyaline vascular type CD. The patient was discharged from our hospital 8 days after the operation without any adjuvant therapy. There was no evidence of recurrence at 1 year after the operation.

DISCUSSION

CD is a rare lymphoid tumor[1] and the etiology is thought to be a hyperplastic lymphatic response related to chronic inflammation with Human Herpes virus 8[6]. CD can be classified into three pathological subtypes; "hyaline vascular type", "plasma cell type" and "mixed type". The hyaline vascular type accounts for approximately 90% of CD, and is usually a solitary tumor characterized by giant lymph follicles with a central vessel and hyalinization. The plasma cell type contains more plasma cells in the tumor, and the mixed type was a few cases[3]. The present case was finally diagnosed as hyaline vascular type CD; however, there

was no evidence for Human Herpes virus 8 infection.

CD occurs at a relatively young age (average 36.6 years) ranging from 8 to 66 years. There is no difference in the frequency between males and females[3]. The clinical characteristics of CD are divided into two groups : "localized type" and "multifocal type". With regard to pathological classification, most of the localized types are the hyaline vascular type, and there are usually no typical symptoms. However, 3% of localized cases can be associated with mass effect symptoms, such as abdominal pain, due to compression of adjacent organs[3,5]. The localized type occurs in much younger patients (mostly in the third decade), whereas the multifocal type is mostly in elderly patients (sixth decade)[5]. Thus, it is likely that localized type tumors in younger patients tend to increase more rapidly than in elderly patients. The suggested treatment for the localized type is resection of the tumor, and surgical treatment is given a good prognosis when resection is completed in an en-bloc manner[2,5,6]. Recurrent tumors of localized CD have only been reported when resection was incomplete. Thus, the five-year survival after operation is nearly 100%[7]. In contrast, in most cases of the multifocal type, it is hard to remove all tumors completely. They are also resistant to other treatment such as corticosteroid therapy, chemotherapy and radiotherapy. Therefore, prognosis of the multifocal type is poor[6,7].

Table 1 summarizes 80 cases of retroperitoneal CD taken from 16 reports[8–23]. Mean age was 41 years ranging from : 11 to 85 years, there appears to be no sexual predominance. Sixty-seven cases (84%) were of the hyaline vascular type, 10 cases (13%) were of the plasma cell type and 3 cases (4%) were of the mixed type. Size of the tumor ranged from 2.5 cm to 14 cm in diameter. Thirty-five cases (54%) were pararenal lesions. These are similar to the cases reported in the

Table 1. Summary of 80 cases of retroperitoneal Castleman's disease in Japan

Sex (M/F) :	36/44	
Mean age :	41 (range : 11–85)	
Histological type :	Hyaline vascular type	67 (83.7%)
	Plasma cell type	10 (12.5%)
	Mixed type	3 (3.6%)
Size (in diameter) :	2.5–14 cm	
Location :	Pararenal	35 (43.8%)

Table 2. Recent cases (16 cases) of retroperitoneal Castleman's disease in Japan

Author	Age	Sex	Location	Size (cm)	Images	Calcification	Preoperative diagnosis	Treatment	Pathological type
Tanaka	12	F	paraadrenal	8	CT, MRI, Ga-scint, MIBG-scint	(−)	adrenal tumor	Res＋LND	HV
Kawakita	56	M	pararenal	4	US, CT, MRI, DIP	(−)	hemangiopericytoma (by biopsy)	Res	HV
Yamamoto	34	F	parapancreatic	6	US, CT, MRI, angio, Ga-scint	(−)	pancreatic tumor	Res＋splenarectomy	HV
Hashimoto	29	F	portal hepatic	5	US, CT, MRI, Angio, Ga-scint	(−)	CD	Res	HV
Kunieda	57	M	pararenal	6	US, CT, Angio,	(−)	retroperitoneal tumor	Res	HV
Shimada	61	F	pararenal	3	DIP, CT, Angio	(−)	retroperitoneal tumor	Res＋nephrectomy＋LND	PC
Furuhata	53	F	pararenal	5	IVP, CT, MRI	(−)	paraganglioma	Res	HV
Ebine	26	F	ileocecal	5	US, CT, MRI, Angio	(−)	leiomyoma, leiomyosarcoma	resection of ileocecum	HV
	45	M	parapancreatic	5	CT, MRI	(−)	pancreatic tumor	Res	PC
Iwamoto	45	F	pararenal	4	DIP, CT, MRI	(−)	ureteral tumor	Nephrectomy＋partial ureterectomy	PC
Kurihara	45	F	pelvis	6	US, CT, Angio	(−)	sarcoma	Res	HV
Asano	52	F	parapancreatic	9	US, CT, MRI	(−)	lymphoma	Res	HV
Kawai	60	F	pararenal	8	US, CT, MRI, FDG-PET	(−)	retroperitoneal malignant tumor	Res＋LND	HV
Kakuta	36	M	pelvis	7	CT, MRI	(＋)	solid tumor	Res	PC
Morimura	50	F	pelvis	4	US, CT, MRI	(＋)	mesenchymal tumor or lymph node	Res	HV
Present case	29	F	pararenal	9	US, CT, MRI	(−)	RCC	nephrectomy＋LND	HV

US : ultrasonography, scint : scintigraphy, angio : angiography, FDG-PET : Fluorodeoxyglucose positron emission tomography, CD : Castleman's disease, RCC : renal cell carcinoma, Res : resection of the tumor, LND : lymph node dissection, HV : hyaline vascular type, PC : plasma cell type.

other countries.

A number of clinical images obtained by ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), angiography, positron emission tomography and Gallium scintigraphy are carried out to make a preoperative diagnosis in Japan. However, the final diagnosis of CD is often made after resection of the tumor, because of lack of specific findings on preoperative image modalities. Generally, CD is shown as a hypoechoic solitary mass by US, which sometimes has calcification. A typical CT image is a solid and homogenous mass with enhancement, and radial or star-shaped calcification might be observed[24]. In addition, on MRI, the tumor shows hypointensity on T1 weighted images and hyperintensity on T2 weighted images. Thus, the differential diagnosis of CD in the retroperitoneal space is often a malignant tumor such as RCC, because of the tumor's hypervascularity.

A recent report suggests that Gallium scintigraphy is useful for diagnosing the hyaline vascular type of CD[25]. In spite of recent advances in radiographic diagnosis, preoperative diagnosis of CD appears to be difficult. Thus, we re-investigated a recent Japanese report of 16 cases of retroperitoneal CD in the last decade (Table 2; from 1996 to 2006). The data showed that 12 cases (75%) were of the hyaline vascular type and 4 cases (25%) were of the plasma cell type. The average diameter of the tumor was 5.6 cm rangeing from 3 to 9 cm. In 7 cases (44%) tumors occurred in the pararenal space. A preoperative diagnosis of suspicion of CD was made in only 1 case using a combination of imaging studies[13] (Table 2). Gallium scintigraphy was performed in only 3 cases and preoperative diagnosis could be made in only 1 case[13]. Calcification was detected in only 2 cases, but the 2 cases were not preoperatively diagnosed as CD. Surgical therapy was done for all cases. In our case, radical nephrectomy and lymphaderectomy were performed based on the preoperative image studies showing RCC characteristics.

In conclusion, these data showed that correct preoperative diagnosis of CD is critically difficult because of the lack of a typical image. The present case suggests that careful and thoughtful consideration is required for these retroperitoneal tumors, especially in younger patients.

CONCLUSION

We have reported a case of pararenal CD, which was indistinguishable from RCC. Based on the preoperative diagnosis of RCC, radical nephrectomy was performed. Pathological findings confirmed the diagnosis of hyaline vascular type CD. The tumor was resected en-bloc with the kidney and there was no recurrence 1 year after the operation.

Regardless of recent progression of radiographic studies, the preoperative diagnosis of CD is still difficult.

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和文抄録

腎癌と鑑別が困難であった後腹膜キャッスルマン病の1例

星　昭夫[1]，新田　正広[1]，徳永　正俊[1]，星野　英章[1,2]
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29歳，女性．健診超音波検査で右腎腫瘍を指摘され当科を受診した．CTでは右腎下極に径9 cm，内部均一で動脈相にて腎実質と同等に造影される腫瘍を認めた．腎細胞癌の術前診断にて根治的腎摘除術およびリンパ節郭清術を施行した．切除標本肉眼所見では腫瘍は薄い被膜を有し腎と完全に隔てられていた．術後病理診断はCastleman病（hyaline vascular type）であった．術後1年を経過した時点で再発は認められていない．

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